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HYDRANENCEPHALY: CASE REPORT AND LITERATURE REVIEW

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ABSTRACT
A case of a one month old girl who was delivered by a teenager and had a normal head size at birth but with a progressive head enlargement is presented. She had normal primitive reflexes at birth. A Computerized tomography showed she had hydranencephaly. A review of the literature on hydranencephaly is also presented.

KEYWORDS: Head Circumference, Hydranencephaly,Neonate, Teenage-mother

CASE PRESENTATION

Baby girl M. D. was delivered by an 18-year-old mother and had a normal head size at birth. However, one week later the head started enlarging and she was then taken to a hospital where ampicillin-cloxacillin combination was prescribed. The head continued to enlarge and she was then taken to anther hospital where she was then abandoned by the mother and subsequently referred to our clinic.

The prenatal and other histories including maternal habits events in the pregnancy could not be obtained.
She was found on examination to have macrocephaly with a head circumference of 51cm at age one month (expected is 35 + or -2cm), the anterior and posterior fontanelles were enlarged and buldging with sutural diasthesis. The primitive reflexes were present but sluggish and there was no sun setting appearance of the eyes. There was no evidence of spinal bifida or lower limb deformity. A diagnosis of Hydrocephalus was made and she then referred to a Neurosurgeon. A C-T scan done showed among other things the presence of only the structures of the posterior fossa, namely, the brain stem, cerebellum and the thalamus which protrude 32cm into the fluid filled cranial cavity. The third ventricle was not obvious and the brain mantle was also not obvious. These are shown in Fig 1. A diagnosis of Hydranencephaly was confirmed and child was sent back to us. She was managed conservatively until her death at age six months.

LITERATURE REVIEW

Hydranencephaly is the complete or near complete absence of the cerebral cortex and basal ganglia, which are then replaced by a membranous sac of fluid, glial tissue and the ependyma in an intact skull. (Pangui *et al* 1991; Byers *et al* 2005) In this situation, some of the primitive reflexes are (Byers *et al* 2005; Kaga 2002). It is thought to follow occlusion of the internal carotid arteries which leads to generalized cerebral infarction. Byers *et al* 2005). Several causes have been advanced as initiators of this occlusion. It could follow an intrauterine viral infection (Parish 1989; Kubo *et al* 1994), especially Herpes simplex (Parish 1989), or it may be metabolic (Castro-Ciago *et al* 1999), oestrogenic (Blare *et al* 1988), genetic or parasitic (Pangui *et al* 1991) or toxic (Nieto *et al*1994). It has even been reported to follow twin-twin transfusion with the recipient developing the condition (Barrent *et al* 2000). A case has been reported to in the vertebral/basilary artery territory (Rossmann , Parks 1978).

Hydranencephaly is generally classified as a circulatory encephalopathy and two opposing hypotheses have been postulated. One is the destructive theory in which the cortex is formed but destroyed in utero and the other is the dysontogenesis in which there is early disruption of organogenesis (Pangui *et al* 1991).

Hydranencephaly can be bilateral or unilateral, in which case only one cerebral hemisphere is involved, leading to hemihydranencephaly (Ulmer *et al* 2005; Greco *et al* 2001). The incidence of bilateral Hydranencephaly is 0.5 per 1000 births (Pangui *et al* 1991), while only about seven cases of hemihydranencephaly have been reported in the medical literature (Ulmer *et al*). The incidence is lower as the maternal age advances. (Lubinsky 1997; Lubinsky *et al* 1997). Our patient's mother was18years.

The diagnosis can be made in utero using ultrasonography or magnetic resonance imaging (Byers *et al* 2005). Postnatally, one can suspect the illness if the child's head size at birth is normal, but increases progressively after birth, with normal primitive reflexes and by Transillumination of the skull (Barozzino , Sgro 2002). CT scan and MRI are important diagnostic tools postnatally (Garcia-Inigo *et al* 2004; Poe , Coleman 1989). The two

major differential diagnoses are extreme Hydrocephalus and bilateral extra cerebral collection of fluid in the skull. These can be differentiated form Hydranencephaly by using an EEG (Guruuaji *et al* 2005; Linuma *et al* 1989). Extreme Hydrocephalus will show evidence of cortical activity while hydranencephaly will not, and will give a flat isoelectric recording (Guruuaji *et al* 2005). There will be no evoked visual potential in hydranencephaly (Linuma *et al* 1989).

Most neurosurgeons believe that surgery is unnecessary as majority will die in infancy (Adeloye 2000). However, some believe that repeated and even complex surgery such as choroid plexectomy can be done (Wellons *et al* 2002). Although most will die in infancy, survival upto 10years have been reported (Corington *et al* 2003). Infact the longest survival reported is twenty years (Corington *et al* 2003). Children with hemihydranencephaly can lead a normal life (Ulmer *et al* 2005)

CONCLUSION

One of the ethical questions is the appropriate treatment for this seemingly fatal illness. Should surgery be offered to them using the merger resources available or wait for the death of these patients whenever it will come. Our patient was referred back to us after the diagnosis was made by C-T scan. Mention must be made that there was a time complex cardiac surgeries were denied children with Trisomy 21. These ethical issues will continue to be considered especially when organ transplantation is involved (McAbee *et al* 2000).

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